Jervell and Lange-Nielsen syndrome in a middle aged patient

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Summary: A case of Jervell and Lange-Nielsen syndrome is reported in a 50 year old woman presenting with repeated ventricular tachycardia terminating fatally. The case is interesting in view of the fact that this syndrome has not previously been reported in a middle-aged patient.

Introduction

Many cases of Jervell and Lange-Nielsen syndrome (JLS) have been recorded since the original report of Jervell and Lange-Nielsen in 1957.¹ This syndrome is essentially a variant of idiopathic long QT syndrome associated with congenital perceptive type deafness and autosomal recessive inheritance. The common mode of presentation is repeated syncopal episodes and/or sudden death related to refractory ventricular tachycardia (VT) which can be polymorphic (torsades de pointes).² A majority of patients present in childhood, though patients presenting in their thirties and early forties have been reported.³ The topic has been fully reviewed by Schwartz *et al.* in 1975³ and recently by several other authors.^{4–8}

This report is presented to highlight the unusual age at presentation of a patient with JLS.

Case report

A 50 year old woman presented with the sudden onset of repeated episodes of loss of consciousness. She was asymptomatic prior to the present illness but was known to be deaf from early infancy. There was no history of sudden death in the family. The patient had no occasion to take any drug which could account for the present illness. Physical examination revealed bilateral perceptive type of deafness. The pulse rate was 70–80/min with frequent extrasystoles. She was mildly hypertensive (blood pressure 150/100 mm Hg). Examination of heart and other systems revealed no abnormality. Electrocardiogram (ECG) revealed a markedly prolonged QTc interval (0.67 seconds) as

calculated by Bazett's formula (Figure 1). There were frequent ventricular premature beats (VPB) with 'R' on 'T' phenomenon and bursts of VT of torsades de pointes variety. Episodes of unconsciousness were documented to be associated with VT. Twelve lead ECG did not reveal any evidence of an acute coronary episode even 2 days after admission. Other routine investigations which included serial serum enzymes and electrolyte estimation were normal. Response to treatment with parenteral propranolol, lignocaine and diphenylhydantoin was unsatisfactory. There was initially some success with drug-induced overdrive suppression (atropine and isoprenaline), but these manoeuvres later on were of no avail. The patient died 3 days after hospitalization. The case was clinically diagnosed as JLS. Routine 12 lead ECG in other family members were found to be normal.

Discussion

The patient presented in her middle age. The diagnosis of JLS was made with reasonable certainty. She had the two most important criteria of JLS. They were congenital perceptive type of deafness and a markedly prolonged QTc interval on ECG. Neither history nor investigation revealed any of the known causes of the latter finding. She succumbed to repeated VT of torsades de pointes variety, a known complication of JLS.

It is difficult to say why she remained trouble free for so long and presented at such an unusual age. Pooled data by Schwartz *et al.*³ show an occasional case presenting above the age of 30, the oldest patient in this review being 40 years old.³ Emotional and physical stress are known to precipitate arrhythmia in this syndrome.⁴ However, a properly timed premature ventricular ectopic might be more important for

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Figure 1 Ventricular tachycardia of torsades de pointes type. A markedly prolonged QTc interval is shown after conversion to normal sinus rhythm (last strip).

induction of ventricular arrhythmias in such patients.⁶ This might explain the long trouble free course in a majority of such patients.

This case thus highlights the fact that JLS can

remain quiescent for a long time. One should not be surprised to encounter a symptomatic patient at relatively late age and for that matter at any age.

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